CASE REPORT

Massive right atrial myxoma presenting as congestive heart failure: an unusual presentation of a rare tumour

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SUMMARY

Cardiac myxomas are the most common type of primary cardiac tumour. The most common location of cardiac myxoma is left atrium. Right atrial myxomas are very rare and usually asymptomatic or sometimes present with dyspnoea. Right atrial myxoma presenting as a right-sided heart failure is very rare. We report a very rare case of a 52-year-old man of right atrial myxoma presented unusually as right-sided heart failure.

BACKGROUND

Metastatic cardiac tumours are more common than primary cardiac tumours. Cardiac myxomas are the most common type of primary cardiac tumour. Its usual site of involvement is left atrium. Unusually it involves the right atrium.¹ The presentation of right atrial myxomas is variable and depends on size, position, mobility of tumour and also on patient body position and physical activity. It can remain asymptomatic or can cause constitutional symptoms as well as dyspnoea.² But presenting chiefly as right-sided heart failure is very rare. Here, we present a very rare case of a 52-year-old man with right atrial myxoma presented unusually as right-sided heart failure.

CASE PRESENTATION

A 52-year-old man, presented with 1-year history of recurrent pedal oedema and abdominal

swelling. There is also a history of fatigue and insidious onset dyspnoea which was initially exertional and gradually progressive from the New York Heart Association (NYHA) class 1 to NYHA class 4 in the last 6 months. Dyspnoea was associated with orthopnoea. He had been also treated with diuretics from his local physician. There was no history of chest pain, syncope or palpitation. He also had a history of fever, malaise and progressive weight loss since the previous 15–18 months.

There was no history of diabetes, hypertension, tuberculosis or asthma. The patient was non-smoker and non-alcoholic.

On physical examination, the patient had tachycardia with pulse rate of 110 bpm and tachypnoea with respiratory rate of 32/min. Blood pressure in right arm supine position was 90/70 mm Hg. Jugular Venous Pressure was raised with prominent a waves and slow y descent moderate ascites and pedal oedema was present. Hepatomegaly was present with liver palpable 4 cm below right costal margin and liver span of 16 cm. Cardiac auscultation reveals mid-diastolic murmur at left third intercostal space and right-sided tumour plop.

INVESTIGATIONS

Routine laboratory investigations showed decreased haemoglobin (11.8 gm/dL), normal total leucocyte count ($10 \times 10^9/L$) and elevated erythrocyte sedimentation rate (40 mm). Total and

Figure 1 (A) A huge RA mass almost completely filling RA cavity and also prolapsing into RV up to greater extent. (B) RA mass is attached to the junction of free wall and roof by a pedicle (shown by thick arrow). RA, right atrial; RV, right ventricle.

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direct bilirubin levels were raised upto 60.4 $\mu\text{mol/L}$ and 45.3 $\mu\text{mol/L}$ (normal: $\leq 17.1 \mu\text{mol/L}$ and $\leq 4.4 \mu\text{mol/L}$, respectively). He was found to be negative for hepatitis B surface antigen and antibody against hepatitis C. Kidney function tests were within normal limits. ECG was normal except sinus tachycardia.

Transthoracic echocardiogram (TTE) showed an echogenic right atrial mass of huge dimension (6.6 $\times$ 4.3 cm), almost completing filling the right atrial cavity (figure 1A). It appeared to be connected to the junction of right atrium free wall and roof by a small pedicle (figure 1B, see online supplementary video 1) and prolapsing through the tricuspid valve into the right ventricle (RV) during diastole and not completely coming back to right atrium with some part remaining in RV during ventricular systole causing obstruction and mild insufficiency of tricuspid valve (see online supplementary video 2). Left ventricle (LV) ejection fraction was 60%. Tricuspid annular plane systolic excursion (TAPSE) was 1.7 cm which denotes normal RV function.

Contrast-enhanced CT chest showed right-sided pleural effusion, bilateral basal segment linear subpleural opacity and large right hypodense filling defect.

After surgical removal, grossly the excised tumour was single, polypoidal gelatinous, friable brown mass measuring 6.6 $\times$ 2.5 cm (figure 2).

Microscopic examination showed typical cytology of myxoma. It showed spindle to fusiform cells in myxoid background. The cells had oval nucleus, bland chromatin and moderate amount of eosinophilic cytoplasm. Occasional multinucleate cells were noted. Stroma showed large areas of haemorrhage focal haemosiderin laden macrophage collection and mild chronic inflammatory infiltrate.

TREATMENT

The treatment of choice for atrial myxoma is a surgical evacuation. In our case, also the mass was surgically removed.

Figure 2 Macroscopic feature of excised tumour showing single, polypoidal gelatinous, friable brown mass. Online supplementary video 1: RA mass is attached to the junction of free wall and roof by a pedicle. Online supplementary video 2: RA mass prolapsing through the tricuspid valve into the RV during diastole and not completely coming back to right atrium with some part remaining in RV during ventricular systole. RA, right atrial; RV, right ventricle.

OUTCOME AND FOLLOW-UP

On close follow-up after surgery, patient had significant improvement in his symptoms.

DISCUSSION

Although the myxoma can present in any age group, but the average age is 50 years.³ This is in accordance with our case, where age of the patient was 52 years. Cardiac myxoma mostly involves the left atrium. Rarely it also involves the right atrium (RA). The presentation of RA myxoma is variable depending on size, position, mobility of tumour and also on the patient's body position and physical activity. It can remain asymptomatic⁴ or can cause constitutional symptoms.⁵ In our case also, the patient had fever, fatigue, anaemia and weight loss (8 kg in 1 year) constituting constitutional signs. These symptoms mostly disappear after surgery. In our case report also, the patient had symptomatic improvement after tumour removal.

RA myxoma can also presents as haemoptysis, dyspnoea, chest pain, syncope if myxoma fragments embolises to pulmonary artery. But presenting chiefly as right-sided heart failure is very rare. To the best of our knowledge, only few cases of RA myxoma presenting as right-sided heart failure has been reported in the literature. In the case report by Ali *et al*,⁶ a 28-year-old man of RA myxoma presented with pedal oedema as a manifestation of right-sided heart failure, but in our case the patient had pedal oedema as well as ascites and hepatomegaly as manifestation of right-sided heart failure. In another case report by Frishman *et al*,⁷ the patient had similar findings but in that RA myxoma was attached to the right atrial free wall as opposed to attachment to the junction of right atrial roof and free wall in our case. In the article by Belle,⁸ two-dimensional echocardiography in all the patients of RA myxoma presenting as right heart failure showed attachment of myxoma to interatrial septum which is the common site of attachment of RA myxoma described in the literature. But in our case, the attachment site was at the junction of roof of right atrium and free wall. TTE is a simple, non-invasive diagnostic tool for imaging RA myxoma. It can tell us about the site of attachment of myxoma whether the tumour is extending to right ventricle or not, embolisation to pulmonary artery, tricuspid stenosis or regurgitation and pulmonary regurgitation.⁹ TTE in our case showed an echogenic right atrial mass of huge dimension (6.6 $\times$ 4.3 cm), almost completely filling right atrial cavity. It appeared to be connected to the junction of right atrial roof and free wall by a small pedicle and

Patient's perspective

I had a big tumour in my right side of heart because of which I had breathlessness and swelling of my whole body. After its removal I am feeling much relieved.

Learning points

- ▶ Although the left atrium is the usual site of involvement of cardiac myxoma, rarely it can also involve the right atrium.
- ▶ RA myxoma also rarely presents with the features of a right-sided heart failure aside from its more common manifestations like dyspnoea, syncope and constitutional symptoms.
- ▶ One of the differential diagnosis of right-sided heart failure, though rare, can be right atrial myxoma.

prolapsing through the tricuspid valve into the right ventricle during diastole and not completely coming back to right atrium with some part remaining in right ventricle during ventricular systole causing obstruction and mild insufficiency of tricuspid valve. Although the LV and RV function was good as denoted by the normal LV ejection fraction and TAPSE, respectively; but because of the huge size of mass which was almost filling whole of RA and considerable portion of RV also, the effective space for venous return and blood filling in the right side of the heart is significantly reduced causing low cardiac output. Also, the mass protruding from RA to RV caused significant valvular obstruction together with insufficiency.

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